Unusual association of diseases/symptoms

A 66-year-old male with lower abdominal lump: an unusual presentation of an uncommon abdominal pathology

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Correspondence to Mr Mohamed Saleem Noormohamed, smithnsal@yahoo.co.uk**Summary**

A 66-year-old Caucasian male was admitted following symptoms of intermittent umbilical pain aggravated after meals and associated with vomiting of contents. Physical examination revealed a tender, partially reducible swelling suspicious of complicated umbilical hernia. Abdominal x-ray revealed dilated small bowel loops which appeared consistent with clinical diagnosis. He underwent a laparotomy subsequently which revealed a small defect in the linea alba with viable small bowel and two firm mesenteric masses encroaching the lumen approximately two feet from the ileocaecal junction. The histology of the excised bowel and masses revealed that the tumour composed of bland spindle cells with slender to plump nuclei and eosinophilic cytoplasm. Mesenteric fibromatosis are the most common primary tumours of the mesentery and constitute about 3.5% of all fibrous tissue tumours. Intra-abdominal desmoids are very rare and benign tumours but are very aggressive and should be considered in the differential diagnosis of acute abdominal pain.

BACKGROUND

Intra-abdominal desmoids are very rare and benign tumours but are very aggressive and should be considered in the differential diagnosis of acute abdominal pain.

CASE PRESENTATION

A 66-year-old Caucasian male presented in August 2008 with a 10 days history of abdominal pain. He described his pain to be intermittent – lasting up to 1–2 h – with multiple episodes daily, located around his umbilicus. He mentioned six episodes of vomiting over the last 3 days, always following food consumption, but no change in bowel habits. Physical examination, revealed a peri-umbilical tender swelling which was partially reducible.

INVESTIGATIONS

Abdominal x-ray showed some dilated small bowel loops centrally.

DIFFERENTIAL DIAGNOSIS

A working diagnosis of strangulated umbilical hernia was made, and a laparotomy was carried out.

TREATMENT

Laparotomy revealed a 3 cm defect of the linea alba containing viable small bowel. About 2 feet proximal to the ileo-caecal junction, two hard mesenteric masses encroaching the lumen partially (each measuring 4–5 cm), were also found. The small bowel containing both masses was removed and side-to-side anastomosis was carried out.

OUTCOME AND FOLLOW-UP

Apart from a postop respiratory infection, the patient had an uneventful recovery and discharged home on the 8th postoperative day.

The histology of the excised bowel and masses was reported, the tumour composed of bland spindle cells with slender to plump nuclei and eosinophilic cytoplasm. They were arranged in long fascicles with collagenous stroma with no obvious mitotic activity. These cells were positive for α -smooth muscle actin and show nuclear positivity for β -catenin staining. They were negative for S100 and MNF-116. The morphological and immunoprofile were those of mesenteric fibromatosis (MF).

The patient had a follow-up CT scan with no residual disease seen in the abdomen or pelvis and no mediastinal abnormality found.

DISCUSSION

The aetiology MF is unknown but usually a secondary response to trauma or hormonal influences (gender, pregnancy and oral contraceptives) or associated with familial adenomatous polyposis (FAP).¹ Nichols in 1923 described the association between MF and FAP.² MF tumours were originally reported in a phenotypic pattern of FAP known as Gardner syndrome which in addition to intestinal adenomatous polyps is also associated with bony abnormalities, ocular fundus pigmented lesions and epidermoid cutaneous cysts and fibromas. Although there is no proven causative relationship in desmoids formation, various mutations of the APC gene have been identified and implicated.

MF can occur in two forms: the sporadic and the secondary form. The sporadic form is rare and is considered to be a variant of a benign stromal neoplasm of fibroblast-myofibroblast origin.

Clinicopathologically desmoid tumours are classified into three types: intra-abdominal, extra-abdominal and abdominal.³ Histologically MF of the abdominal wall and extra-abdominal MF (desmoids outside the abdominal wall) are similar. Intra-abdominal desmoids are often invasive and metastasise to areas surrounding neurovascular

structures; with higher recurrence rates after surgery. Trauma was considered as one of the major causative factor for the onset of this type of tumour.⁴

Among other causative factors accidental blunt injuries, lacerations, intramuscular injections, fractures, and different endocrine or genetic factors have also often been implicated.⁵

Abdominal USG and CT are the most useful imaging techniques for determining the exact localisation of the tumour which is usually, a solid lesion observed as a mass with soft tissue density.⁶ Mesenteric tumours are more common in women in their reproductive years; especially during pregnancy and postpartum.⁷ Menopause and menarche has been associated with tumour regression.^{7 8}

The cytosol oestrogen receptor binding characteristics and the incidence was investigated by Lim *et al*, found that oestrogen receptors were present in 33% of the desmoid tumours.^{9 10} MF more commonly presents itself as large palpable tumours resulting in abdominal pain otherwise the signs and symptoms are subtle. The other documented symptoms in literature were intestinal and ureteral obstruction, weight loss and mesenteric ischaemia. Mechanical intestinal obstruction is extremely rare. The differential diagnosis includes gastrointestinal stromal tumours, lymphomas, carcinoid tumours, fibrosarcomas or inflammatory fibroid polyps.¹¹

Multiple treatment were described in the literature, ranging from wide surgical excision, cytotoxic chemo radiotherapy, non-steroidal anti-inflammatory drugs and antioestrogens, outcome from these are unpredictable and controversial. The primary goals of the operations are to preserve function and structure, as the presence of residual disease not clearly linked to show any benefit on 5-year disease free or overall survival. Surgical wide excision of the tumour is the recommended treatment of choice, as these tumours have a tendency toward local recurrence.^{4 6 7 11–15} Despite radical surgery recurrence rates in most studies can be in the range of 25–50%. Achieving negative resection margins may not prevent local recurrence and may result in unnecessary morbidity.¹⁶ In addition, Kollevold *et al* have reported temporary tumour regression following salpingo-oophorectomy in a patient with breast cancer.¹⁷ Whereas, Baliski *et al* proposed that delayed surgery with neoadjuvant chemo radiotherapy treatment is a better option than surgery excision alone.¹⁶ The outcome following radiotherapy was not obvious, but several reports advocated that using dose levels greater than 50 Gy achieved complete regression of tumours. Nuytens *et al* reported a significantly lower local recurrence rate following radiotherapy or surgery with radiotherapy.^{6 17 18} The patient presented in this report had an acute abdominal pain and underwent only an aggressive surgical intervention. At follow-up at 33 months the patient was still disease free. As a result, we conclude saying that intra-abdominal

desmoids are rare aggressive, but benign tumours and carefully considered in the differential diagnosis of acute abdominal pain.

Learning points

- Intra-abdominal desmoids are very rare and benign tumours but are very aggressive and should be considered in the differential diagnosis of acute abdominal pain.
- Abdominal USG and CT are the most useful imaging techniques.
- Surgical wide excision of the tumour is the recommended treatment of choice.
- Local recurrence.

Competing interests None.

Patient consent Obtained.

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